



Case report

Sudden death due to Dysembryoplastic Neuroepithelial Tumor

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ABSTRACT

Sudden death in adults from presumably natural causes occurs more frequently than is commonly thought. Numerically they constitute a significant fraction of the total mortality of which central nervous system causes account for 15 percent of the cases. The neoplasms of neuroepithelial origin account for 3 percent of these cases. We are presenting a case of 40 year old woman, who suddenly developed vomiting, abdominal pain and rapidly deteriorating vitals over a period of 20 h and breathing last on the way to the hospital. A cystic swelling of the right frontal lobe was present at autopsy, histopathological examination of the swelling revealed to be Dysembryoplastic Neuroepithelial Tumour (DNT). DNT is a rare neurological tumor characterized by presence of neurons, astrocytes and oligodendrocytes presenting with complex partial seizures. It frequently affects the frontal and temporal lobes of adolescents and young adults.

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1. Introduction

Dysembryoplastic neuroepithelial tumor is a recently recognized neuroglial tumor in the world of neurology.¹ DNT are supratentorial tumors of the brain, two thirds of these tumors arise in the temporal lobe and one third in the frontal lobe. The most common symptom is the occurrence of temporal lobe seizures with focal neurological deficits. They are usually reported in the second or third decade of life. Sudden deaths due to Dysembryoplastic neuroepithelial tumour have not been reported till date. In cases of sudden, unexpected deaths of an apparently healthy individual, the role of the Forensic Pathologist comes to the forefront to arrive at a cause and manner of death. We are reporting a rare case of sudden death due to DNT.

2. Case report

A 40 year old woman with symptoms of dizziness and weakness of 4 months duration suddenly developed nausea, headache and vomiting and died on the way to hospital. An autopsy was requested by the law enforcing authorities to ascertain the cause of death. At

autopsy thoracic and abdominal cavity did not show any significant findings. The brain weighed 1400 g and showed the following salient features; a nodular cystic swelling measuring 6 × 5 cm situated in the right frontal lobe with loss of sulci and gyri (Fig. 1). The entire brain was softened and edematous with bilateral tonsillar herniation and midline shift. No other significant findings were present. Viscera were subjected to chemical analysis for determination of the presence of any toxic compound which was negative for the same.

Histopathological examination of the brain lesion showed multiple cystic spaces with proliferation of oligodendroglial cells (Fig. 2), which was suggestive of Dysembryoplastic neuroepithelial tumor.

The cause of death was opined as death due to Brain tumor (Dysembryoplastic Neuroepithelial Tumor).

3. Discussion

Dysembryoplastic neuroepithelial tumor (corresponding to WHO grade I) is a newly recognized brain lesion first reported in 1988 by Daumas-Duport et al.¹ The authors described five cases of DNT, which occurred in young people and were characterized by partial seizures. DNT is a benign tumor that can be cured by surgical excision^{2,3} and is a cause of partial epilepsy in young adults.³ They usually present with symptoms and signs of increased intracranial pressure rather than focal neurological impairment.⁴ The tumor is differentiated into Simple, complex and nonspecific types.⁵

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Fig. 1. Tumour involving the right frontal lobe.

The exact prevalence of DNT is not known; thus far 200–300 cases have been reported in the Western literature, but none in Indian literature. There is slight preponderance for male sex with age of onset ranging from 1 week⁶ to 60.5 years.⁷ DNT's are usually located supratentorially within the cortical regions of the hemispheres, but may also be located infratentorially and most commonly they are situated in the temporal lobe, either involving the convexity, the anterior portion or abutting the mesial structures (or a combination of these).⁵ Frontal lobe DNT's are second most common and shows a prevalence of 30%, followed by parietal and occipital lobes that account for 10% of all the cases.¹ The signs and symptoms are related to the location of the DNT with a vast majority of patients presenting at an early age (9–10 years) with seizures. The seizures are of partial complex type with or without secondary generalization, followed by simple partial seizures. Occasionally symptoms related to the presence of an intracranial space-occupying lesion, such as headache and vomiting which are the sentinel manifestations of intracranial pathology. In the latter case the duration of symptoms may be short, but it may be protracted up to 60 years.⁸ Radical resection is considered to be the sole treatment of choice in DNT's. Follow up period of 10 years in case of simple form and complex form; whereas follow up to 22 years is required for nonspecific type.^{7,9}

Histologically, the tumor is characterized by multinodularity, predominantly cortical based, although focal extension into the underlying matter may be seen. Predominant cell type is the oligodendrocyte which are atypically arranged against a focally microcystic background.¹⁰

In the present case the deceased was aged 40 year without any previous history of seizures or other symptoms suggestive of DNT. The duration of symptoms of headache and vomiting lasted for less than 24 h and the condition deteriorated rapidly culminating in a sudden and unexpected death of an apparently hale and healthy woman, raising suspicion as to the cause and manner of death. An autopsy with pathological diagnosis of the brain tumor alleviated all the apprehensions of both, the relatives as well as the law enforcement authorities.

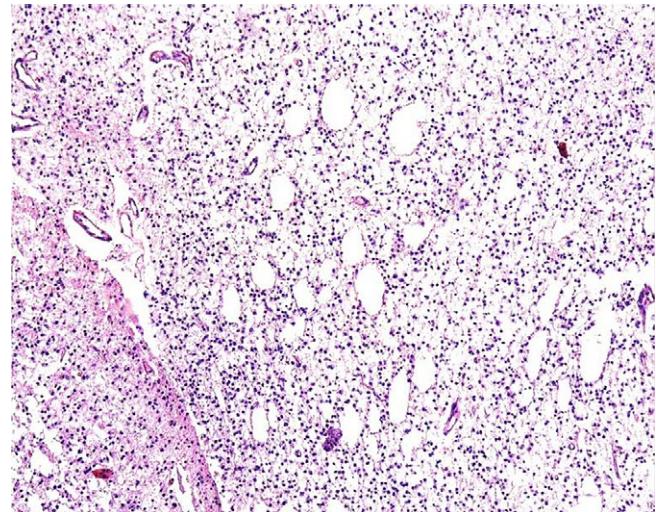


Fig. 2. Microscopic view showing multiple cystic spaces surrounded by oligodendrocytes (10X).

Conflict of interest

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Ethical approval

None declared.

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